Don't trust a vein graft to treat carotid aneurysm in patients with Behçet disease

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Extracranial carotid aneurysm is a rare vascular manifestation of Behçet disease. To our knowledge, only 32 cases have been reported. This article presents a complex case of a 28-year-old man who was first treated by vein graft reconstruction. At 12 months of follow-up, a nonanastomotic false aneurysm of the vein graft occurred and was treated by interposition of prosthetic graft. Two months later, an anastomotic pseudoaneurysm between the two grafts was excluded by two stent grafts. Based on our experience and a review of the literature, we compared the outcomes of prosthetic and autologous vein reconstructions and discussed the role of carotid ligation and immunosuppressive treatment. (J Vasc Surg 2010;52: 471-4.)

Behçet disease is a rare systemic vasculitis, characterized by recurrent oral and genital aphthae, skin lesions, and ocular manifestations. Vascular manifestations are common, with venous thrombophlebitis being most frequent, followed by arterial aneurysm and occlusion. Aneurysms occur preferentially in the abdominal aorta and pulmonary arteries.[1] To our knowledge, only 32 cases of extracranial carotid aneurysm have been reported.[2-21] Since the main vascular manifestation of Behçet disease is thrombophlebitis, prosthetic grafts[8-14] are preferred to autologous vein grafts[5,15,17] for treatment of carotid aneurysm. This article describes a complex case treated by vein graft reconstruction but requiring several reoperations. Based on our experience and a review of the literature, we compared the outcomes of both reconstructions and discussed the role of carotid ligation and immunosuppressive treatment.

CASE REPORT

A 28-year-old West Indian man was admitted for a mass ongoing in the neck for 3 weeks. He had a 2-year history of recurrent oral aphthae and folliculitis. Physical examination revealed a pulsating mass in the right cervical region with severe folliculitis of the overlying skin. The patient's temperature was 38°C. Neurologic and ophthalmic examination revealed no abnormality. Blood tests showed a C-reactive protein level at 28.7 mg/dL (normal, <5 mg/L) with white blood cell count of 13.2/μL. Computed tomography (CT) scan depicted a contained rupture of a 4 cm sacciform aneurysm in the right common carotid artery (CCA) immediately proximal to the carotid bifurcation.

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Surgery was performed immediately to prevent rupture. The CCA and internal carotid artery (ICA) were controlled and clamped. The aneurysm was then opened, and the external carotid artery (ECA) was occluded using a balloon catheter. Because of folliculitis, continuity between the CCA and ICA was restored by interposing an apparently healthy saphenous vein harvested from the thigh. The ECA was ligated. No postoperative complications occurred, and the patient was discharged on the 10th postoperative day.

Examination of biopsy specimens collected from the carotid and surrounding zones showed no infection, but did show destruction of the media and intima due to lymphocyte infiltration. Tests for HLA B51, autoimmune antibodies (serum antinuclear, anticardiolipin, and antineutrophil cytoplasmic antibodies), and infectious disease (human deficiency virus, hepatitis B and C virus, and syphilis) were negative. Based on case history, the diagnosis of Behçet disease was made. The patient was prescribed aspirin 160 mg/day and colchicine 1 mg/day. Ultrasonography at 6 months showed a patent graft and the patient remained symptom-free for 1 year.

Twelve months after intervention, the patient was referred to our institution for right cervical pain. Physical examination showed no abnormality except swelling in the right cervical region and oral aphtae. CT scan revealed a nonanastomotic 3 cm vein graft pseudoaneurysm (Fig 1). The patient was rapidly transferred to the operating room. The dissection of the distal anstomosis between the ICA and vein graft was difficult due to inflammatory tissue. After cross-clamping the CCA and vein graft distally, the pseudoaneurysm was opened, and a defect 1 cm in diameter was identified in the vein wall (Fig 2). A polytetrafluroethylene (PTFE) graft was implanted between the CCA and healthy distal part vein graft. Microscopic examination of the vein showed intima and media destruction. The patient was discharged without complications and prednisone 20 mg/day was associated with colchicine and aspirin. Routine clinical and ultrasonography findings at 2 months were normal.

Three months after reoperation, the patient was readmitted for a pulsatile right cervical mass that developed suddenly after sneezing. At that time, oral aphtae reappeared for 5 days. Just before iodine injection for CT scan, the mass suddenly expanded, and blood pressure dropped. Emergency endovascular exclusion

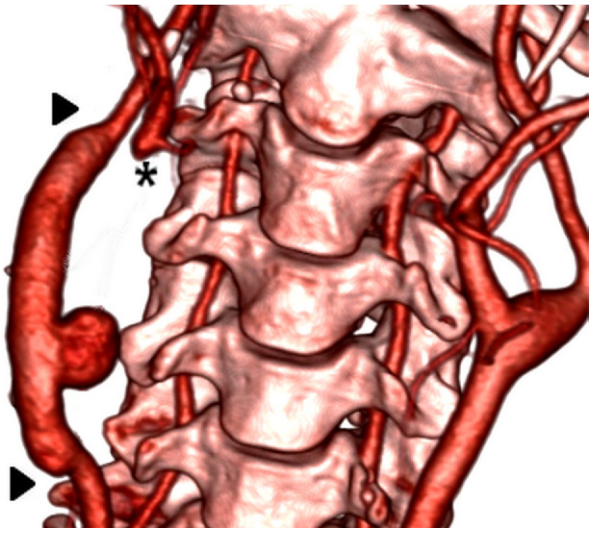

Fig 1. Computed tomography scan of the ruptured vein graft. Arrowheads indicate proximal and distal ends of the vein graft. Asterix shows external carotid artery previously ligated.

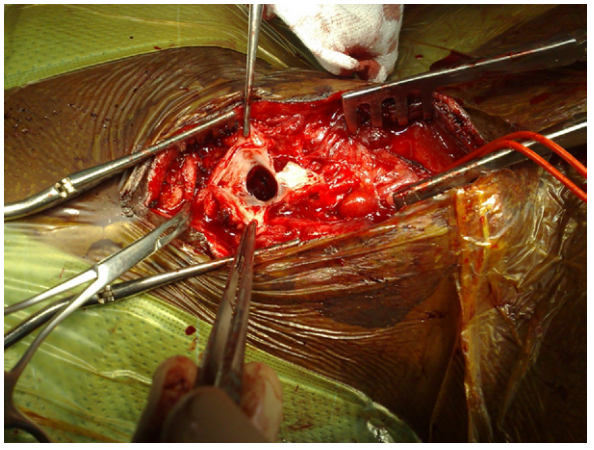

Fig 2. Operative view of vein graft rupture after vessel loop control of common carotid artery (CCA) proximally and clamp control of the vein graft distally.

was decided after evaluation of surgical risks (massive hematoma, second reoperation). Selective angiography via a femoral access (Fig 3) revealed a pseudo-aneurysm at the anastomosis between the remaining vein graft and PTFE graft. A 0.25 Terumo guidewire was placed distally in the ICA. Overlapping Hemobahn (W. L. Gore, Flagstaff, Ariz) stent-grafts (distal, 5 cm × 9 mm; proximal, 5 cm × 11 mm) were deployed in the remaining vein graft (diameter, 10 mm), so as to cover the proximal end of the ICA and distal end of the PTFE graft, respectively. Completion arteriography (Fig 3) showed complete exclusion of the pseudo-aneurysm and a patent ICA. Postoperative recovery was uneventful. Prior to discharge on day 9, CT scan showed stent graft occlusion but no ischemic brain lesions. Since neurologic examination was normal, the patient was discharged. Prescribed medication included aspirin

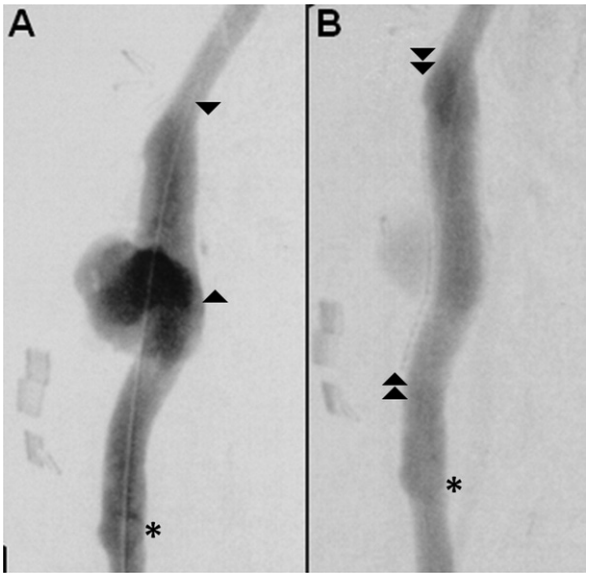

Fig 3. A: Diagnostic angiogram showing pseudoaneurysm at anastomosis between the PTFE and vein graft. **B:** Completion angiogram after deployment of two overlapping stent grafts. *, anastomosis between the common carotid artery and PTFE graft; single arrowheads, proximal and distal ends of the remaining vein graft; double arrowheads, proximal and distal ends of the stent grafts.

160 mg/day, colchicine 1 mg/day, prednisone 80 mg/day, and azathioprine 150 mg/day. Twelve months after the stenting, the patient was asymptomatic, and ultrasonography showed no aneurysm at other locations.

DISCUSSION

Since the main vascular manifestation of Behçet disease is thrombophlebitis, prosthetic grafts are preferred to autogenous vein for treatment of carotid aneurysm (Table I). Only one[9] out of eight prosthetic grafts[8-14] occluded, but three out of six autologous venous reconstructions,[5,15,17] including our case, failed.[5,17] Despite suspicion of Behçet disease in our case, we decided against use of prosthetic material due to presence of folliculitis. To our knowledge, this is the first report describing venous graft rupture in a nonanastomotic site. This suggests that vasculitis can involve venous tissue and supports preferential use of prosthetic grafts.[13,21] Suturing with wide bites in vessel segments away from the diseased artery may lower the risk of pseudoaneurysm after both prosthetic and autogenous reconstruction.[22] In our patient, dissection of the distal vein graft during the first reoperation was difficult, but we identified a presumably "healthy" segment for anastomosis of the PTFE graft. The presence of inflammation should have led us to suspect a diseased vein wall. It is also for this reason that simple patch closure of the arterial wall defect[7] is not recommended.

As suggested previously,[16-20] management of aneurysm in Behçet disease may benefit from endovascular techniques. Indeed, stenting would eliminate the risk of anastomotic

Table I. Clinical findings in case reported herein and 24 previously reported cases of vascular reconstruction to treat extracranial carotid aneurysm due to Behçet disease

Study	*Age (y)/ Gender*	*Localization*	*Treatment*	*Preop medic*	*Postop medic*	*Status (Follow-up)*
Park (2)	25/F	Bilateral CCA	Not described	?	?	?
Canova (3)	32/M	ICA	Not described	?	?	Alive and well (11 mo)
Dhobb (4)	44/F	ICA	Resection and end-to-end anastomosis	?	?	Patent (12 mo)
Sayed (5)	21/M	CCA	Resection and end-to-end anastomosis	S + CPP	CPP + CC	Died from ruptured pulmonary aneurysm (2 mo)
Sayed (5)	30/M	CCA	Direct suture	S + CPP	CPP + CC	Alive (?)
Antar (6)	13/M	CCA	1. Direct suture	?	?	Pseudoaneurysm? (3 mo)
			2. Ligation	?	S + CHB	Alive (48 mo), AAA reported
Sasaki (7)	16/M	CCA	1. PTFE patch	?	?	Pseudoaneurysm (2 mo) alive
			2. Ligation	?	?	(3 mo)
Suzuki (8)	16/M	CCA	PTFE bypass	No	S	Patent (24 mo)
Tacal (9)	35/M	ICA	Tube grafting	?	?	Occluded (2 weeks)
Kuzu (10)	?	?	PTFE bypass	?	?	Alive (?)
Tuzuner (11)	16/M	CCA	PTFE bypass	CC + S	CC	Patent (84 mo)
Özyazıcıoglu (12)	34/F	CCA (iatrogenic puncture)	PTFE bypass	No	AZT + S	?
Iscan (13)	43/M	ICA	PTFE bypass	?	?	Patent (35 mo)
Bouarhroum (14)	26/M	CCA	PTFE bypass	No	S	Patent (12 mo)
Bouarhroum (14)	41/M	CCA	PTFE bypass	No	S	Patent (11 mo)
Posacioglu (15)	31/M	CCA	Vein graft	No	Not detailed	Patent (16 mo)
Sayed (5)	31/M	Bifurcation	Vein graft	S + CPP	CPP + CC	Patent (?)
Sayed (5)	35/M	ICA	1. Vein graft	S + CPP	CPP + CC	Pseudoaneurysm (6 mo)
			2. Ligation	CC	CPP + CC	Alive but minor stroke (?)
Sayed (5)	30/M	ICA	Vein graft	S + CPP	CPP + CC	Patent (?)
Bonnotte (16)	47/M	ICA	Bare stent, coil embolization	CC	AZT	No complication (48 mo)
Park (17)	32/M	CCA (previously vein grafted)	Stent graft (Jostent, Jomed)	?	?	Occluded (6 mo)
Kwon Koo (18)	32/M	CCA	Stent graft (Jostent, Jomed)	?	S + AZT	Occluded (41 mo)
Caballol (19)	29/M	ICA	Stent graft (Wallgraft, Boston)	S + AZT	S + AZT	Patent (27 mo)
Ohshima (20)	56/M	ICA	Stent-graft (Passager, Boston) coil embolization	?	S + AZT	Patent (12 mo)
Berard et al (current study)	28/M	CCA	Vein graft	No	CC	Ruptured (12 mo)
			PTFE graft	CC	S + CC	Pseudoaneurysm (3 mo)
			Stentgraft	S + CC	S + CC + AZT	Occlusion (9 days)

AAA, Abdominal aortic aneurysm; *AZT,* azathioprine; *CC,* colchicine; *CCA,* common carotid artery; *CHB,* chlorambucil; *CPP,* cyclophosphamide; *ICA,* internal carotid artery; *mo,* month; *PTFE,* polytetrafluoroethylene; *S,* steroids.

pseudoaneurysm, provided that a healthy arterial landing zone is available. Occlusion occurred in two of the four reported cases of endovascular treatment using poorly-flexible first-generation balloon-expandable covered stent grafts.[17,18] In our patient, because of the hazardous proximal control due to massive hematoma at the base of the neck, and the impossibility of dissecting the distal part of the vein graft during previous reoperation, we preferred endovascular treatment to ligation. The 11-mm proximal stent graft was oversized in relation to the 8-mm PTFE graft. However, we had already used our last 9-mm stent graft distally. This problem illustrates the difficulties associated with emergency treatment (ie, insufficient time for sizing based on CT scan reconstruction and for ordering appropriately sized stent grafts). Asymptomatic occlusion of our stent graft suggests that particular attention is necessary with regard to carotid artery ligation. However, postoperative follow-up in three previous cases [6,7,21] and in an eight-patient report[5] involving ligation (Tables I and II) were uneventful, except in one case of secondary ligation to treat pseudoaneurysm complicating primary vein grafting.[5] In their series, Sayed et al[5] proposed ligation if stump pressure >70 mm Hg and demonstrated that this option appears to be a reasonable alterative in high-risk surgical patients.

Medical treatment is a keystone for management of vascular manifestations of Behçet disease.[22,23] In our case, occurrence of acute oral ulcerations in all three management episodes suggests that medical treatment was ineffective. Vein rupture and anastomotic pseudo-aneurysm occurred despite therapy using colchicine alone and colchicine plus corticosteroids, respectively. The European League Against Rhumatism recommendations suggest that immunosuppressors lower the recurrence risk and propose cyclophosphamide and cortico-

Table II. Clinical findings in eight previously reported cases of primary ligation to treat extracranial carotid aneurysm due to Behçet disease

Study	*Age (y)/Gender*	*Localization*	*Treatment*	*Preop medic*	*Postop medic*	*Status (Follow-up)*
Tuzun (21)	25/M	CCA (ruptured)	Primary ligation	?	CPP	Alive (1 mo)
Sayed (5)	37/F	ICA	Primary ligation	S + CPP	CPP + CC	Died from cardiomyopathy (4 mo)
Sayed (5)	25/M	ICA	Primary ligation	S + CPP	CPP + CC	Alive (?)
Sayed (5)	25/M	Bifurcation	Primary ligation	S + CPP	CPP + CC	Alive (?)
Sayed (5)	33/M	ICA	Primary ligation	S + CPP	CPP + CC	Alive (?)
Sayed (5)	32/M	ICA (ruptured)	Primary ligation	No	CPP + CC	Alive (?)
Sayed (5)	25/M	ICA	Primary ligation	S + CPP	CPP + CC	Alive (?)
Sayed (5)	37/M	Bifurcation	Primary ligation	S + CPP	CPP + CC	Alive (?)

CC, Colchicine; *CCA,* common carotid artery; *CPP,* cyclophosphamide; *ICA,* internal carotid artery; *mo,* month; *S,* steroids.

steroids for management of peripheral arterial aneurysms, but the optimal dose and duration of the therapy are still in question.[23] As proposed by many authors (Tables I and II), preoperative immunosuppression should be initiated immediately, but postponing the intervention is not always feasible, especially in symptomatic aneurysms.[5,13]

In summary, analysis of the literature and our report has four main implications for treatment of carotid aneurysm in BD. First, synthetic grafts should be preferred to autogenous grafts. Second, endovascular exclusion based on careful CT scan vessel sizing appears to be a promising option, but further study is needed to assess long-term benefits. Third, ligature should be considered in case of intraoperative difficulty. Fourth, aggressive postoperative immunosuppressive therapy is necessary to control disease activity.

AUTHOR CONTRIBUTIONS

Conception and design: XB, DM
Analysis and interpretation: XB, J-MC, HT, GS, VB, DM
Data collection: XB, HT, GS
Writing the article: XB, J-MC
Critical revision of the article: XB, J-MC, HT, GS, VB, DM
Final approval of the article: XB, J-MC, HT, GS, VB, DM
Statistical analysis: N/A
Obtained funding: N/A
Overall responsibility: XB

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